Agenesis of dorsal pancreas with eventration of diaphragm and intrapancreatic pseudocyst: a rare entity

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Abstract

A case of acute pancreatitis in a 43-year-old male patient, which was eventually diagnosed as agenesis of dorsal pancreas with eventration of left hemidiaphragm and intrapancreatic pseudocyst is being reported.

Introduction

Agenesis of dorsal pancreas with other associated congenital malformations is a rare entity.[1] In most of the cases it is associated with other medical conditions, commonest being hyperglycemia, demonstrated in 50% of cases.[2] Only a few cases of dorsal agenesis of pancreas with other associated congenital malformations have been reported in the literature, and its association with other anomaly and disease process may have resulted in more severe clinical manifestations and would have complicated the presentation of this entity. To the best of our knowledge, this was the first case of dorsal pancreatic agenesis occurring in association with eventration of diaphragm.

Case Report

A 43-year-old chronic alcoholic male patient presented in emergency with complaints of severe diffuse pain abdomen associated with vomiting of 2 h duration with a history of mild upper abdominal pain for last 15 days. He was not a known diabetic or hypertensive. On clinical examination, the patient had diffuse tenderness and distension of abdomen and no guarding or rebound tenderness. Laboratory investigations revealed leucocytosis with counts of 16,500, raised serum amylase and lipase with normal liver enzymes and alkaline phosphatase and random blood sugar was raised. X-ray chest and abdomen showed raised left hemidiaphragm and no air fluid levels or free gas under diaphragm. Ultrasonography of the abdomen did not reveal any significant abnormality. Diagnosis of acute pancreatitis was made and patient was managed conservatively and responded well to treatment but his blood sugar level remained high for which he was given insulin. Contrast enhanced computed tomography (CECT) of abdomen was performed after 6 days of admission, which showed intrapancreatic pseudocyst in the head of pancreas with dorsal agenesis of pancreas and eventration of left hemidiaphragm (Figures 1 and 2). Patient was discharged in stable condition after 9 days of admission on oral antibiotics and insulin.

Discussion

Herophilus of Chalcedon (334-280 BC) first noted the existence of the pancreas and Galen (131-200 AD) first described the pancreas as glandular and identified its arterial and venous supply.[3] At embryogenesis, the pancreas develops from the endoderm - lined dorsal and ventral buds of the foregut. The inferior part of the head and the uncinate process of pancreas are formed from the ventral pancreatic bud; the superior part of the head, the neck, the body, and the tail of the pancreas are formed from the dorsal pancreatic bud.[4] Agenesis of dorsal pancreas is a rare anomaly and may be discovered in the course of an operation.[5] If isolated there may not be any symptom or may present with abdominal pain, a medical history of diabetes or may be discovered due to the presence of other anomalies along with agenesis of dorsal pancreas or because of the inflammation of pancreas like in our case. There are case reports of dorsal agenesis of pancreas occur-

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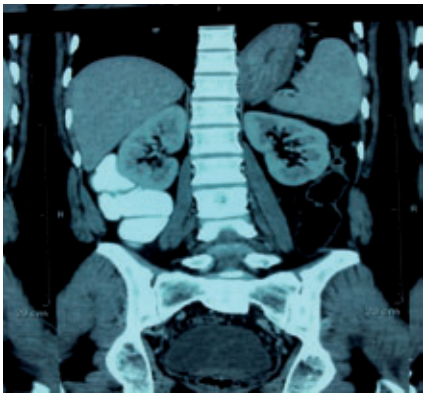

Figure 1. Contrast enhanced computed tomography of abdomen showing high up left hemidiaphragm.

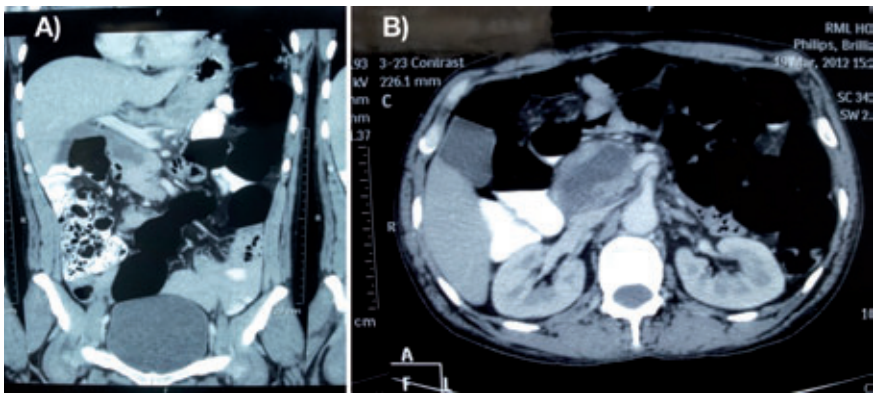

Figure 2. Contrast enhanced computed tomography of abdomen showing agenesis of dorsal pancreas (A) with intrapancreatic pseudocyst (B).

ring in association with other anomalies such as horse shoe kidney and hydrocephalous,[1] polysplenia,[6] retroaortic left renal vein[7] but in this case agenesis was associated with eventration of diaphragm and the patient presented due to the inflammation of pancreas and pseudocyst formation while he was unaware of his diabetic status. Patient may present with acute onset abdominal pain without any evidence of pancreatic inflammation which could be due to insufficient drainage of pancreatic duct system but in our case the cause of pain was pancreatitis[8,9] and pseudocyst formation. Diagnosis was made by CECT scan[10] and it is important to rule out other anomalies of pancreas, atrophy of pancreas due to chronic recurrent pancreatitis and also in cases of carcinoma of head of pancreas[11,12] where Whipple's procedure needs to be done and there may not be any remnant pancreas for anastomosis. The importance of diagnosis lies in the potential clinical consequences of agenesis of dorsal pancreas and associated eventration of hemidiaphragm.

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